Cochlear implantation in adults with early-onset deafness

Chapter 7

General discussion and valorisation

The aim of the current thesis was to expand current knowledge regarding cochlear implantation in a very specific patient group: adults or adolescents with an onset of deafness in early childhood. In the different chapters of this thesis we presented data on the outcomes of this patient group with CI and on the factors that might influence these outcomes, either patient-related or related to the fitting and coding of the electrical signal.

Defining the target group

One of the difficulties we came across in the course of this thesis was how to define and demarcate our specific target group of patients. In the different chapters they are referred to as congenitally, prelingually, perilingually, as well as early-deaf(ened) patients. Where the first three terms each refer to a different age at onset, “early” is a more comprehensive term, generally referring to any age at onset before the end of the language acquisition period. Although the term “prelingual deafness” has been used quite broadly in literature to indicate an onset before the age of 4 or even 6 years (Santarelli et al., 2008; Teoh et al., 2004a; van Dijkhuizen et al., 2016), and has also been used in that sense in *chapters 2 and 3* of this thesis, it is actually defined as an onset before the beginning of the language acquisition period.

The differentiation of patients based on age at onset is important, as it is presumed that even minimal auditory exposure affects auditory development compared to no auditory exposure at all (Kral, 2007), but at the same time it is practically unfeasible as the medical and audiological information required to make such a differentiation is usually lacking, especially in patients born before the introduction of new born hearing screening. Even the clinical classification of a patient as “early” deaf, the term we adopted in *chapters 4, 5 and 6*, is mostly based on the combined information obtained from a number of sources such as available medical files and audiograms from the past, presence of deaf speech, type of education received, and recall of family members on the onset of the hearing impairment.

An important aspect adding further variability to our target group is that not only the age, but also the degree and configuration of the hearing loss are often unknown for patients born several decades ago. Taken together, our target group of early-deafened patients is necessarily diverse, the use of strict definitions being unfeasible, primarily due to the lack of sufficient information on the hearing history of many patients.

From another angle, the demarcation of our target group is equally determined by the age at which cochlear implantation is performed. The main issue is whether subjects implanted as adolescents or adults, with a comparable onset of deafness, can be seen as one group or not. From what implantation age onwards is it reasonable to say that the influence of the early onset of the hearing impairment outweighs the influence of the age at implantation? Is there for example a difference between early-deafened subjects implanted at age 12 vs. 25, based on their age at implantation? In the review in *chapter 5*, we concluded that age at implantation is very likely not related to CI outcomes in adults with early onset of deafness, whereas in adolescents this relation does need further investigation. This implies that, until this is further clarified, the better option would be to separately analyse outcomes of early-deafened adolescents and adults if the aim is to see how these subjects perform as a group. This is also the case in *chapters 2, 3 and 6* of this thesis, where all subjects were implanted after the age of 18 years. On the other hand, based on evidence from N1 and P1 potentials, Sharma et al. (2015) showed that “in all likelihood, the sensitive period ends by age 7 years, resulting in a re-organized auditory cortex that is unable to effectively process the stimulation provided by the cochlear implant”. The reasons why the outcome of CI would be different for adolescents and adults, as sensitive periods are closed in both cases, is an additional question that would then have to be addressed. Potentially other factors such as education and early hearing aid rehabilitation could explain the difference, rather than the younger age itself.

Optimizing fitting parameters

C*hapters 2 and 3* of this thesis revolve around how technical aspects of cochlear implant signal processing, such as fitting parameters and properties of the CI signal, might affect outcomes in our target group of CI users. Current cochlear implant processing generally employs a “one size fits all” approach with respect to most fitting parameters, such as stimulation rate, number of stimulated electrodes, frequency allocation, etc. Although these default settings work well for most postlingual CI users, they are not necessarily optimal for early-deafened CI users who have developed a completely different auditory system due to their early onset of auditory deprivation. We therefore hypothesized that a more individualised approach towards fitting could improve performance, with psychophysical measures of certain signal processing aspects being used to change fitting parameters.

In *chapter 2* we measured amplitude modulation detection thresholds (AMDTs) to assess temporal processing abilities of both pre- and postlingually deafened CI users. This method is commonly used since the CI processor equally applies amplitude modulations to the envelope of the fast electrical pulse trains that are delivered to the electrodes. We were able to demonstrate that the amplitude modulation detection abilities of the early-deafened group were significantly poorer than those of the postlingually deafened group, especially for fast modulations. In addition, AMDTs were significantly correlated with speech recognition measures, also for the early-deafened subjects. If this relation were to be a causal one, a better detection of amplitude variations over time could potentially lead to better speech understanding. Consequently, it would be interesting to investigate whether improving the transmission of these temporal cues by means of changes to the speech processing strategy, could lead to improved speech recognition. So far, however, research efforts focusing on the effects of changing stimulation rate and/or stimulation level, which both influence the transmission of temporal cues, have not been successful at finding consistent and significant improvements on temporal or speech processing abilities (overview in Brochier, McDermott, and McKay (2017)). Since these studies are generally performed in postlingually deafened CI users, and we found that temporal processing was significantly poorer for the average early-deafened CI user, it would be interesting for future studies to investigate these relations specifically in an early-deafened subject group. In addition, as our study showed that the ability to detect slow modulations was related to speech understanding performance in this group, those subjects having most difficulty detecting these slow modulations are also the ones for which it would be most interesting to investigate the effect of changing these parameters.

Where *chapter 2* focuses on the processing of temporal cues, *chapter 3* gives attention to the coding of spectral parameters, related to the place of stimulation. In *chapter 3* we first measured spectral processing through the ability to perform electrode discrimination, and then attempted to optimize the processing of spectral cues by means of deactivating all non-discriminable electrodes. A clear advantage of the adapted fitting on outcomes could not be found, however. In addition to the limited number of subjects, the influence of a number of other parameters that were changed along with the deactivation of electrodes could not be ruled out, and gives rise to further investigation.

In *chapter 3*, the limited number of subjects unfortunately did not allow for an investigation of the relation between spectral discrimination performance and speech understanding outcomes with CI, but other studies have been able to demonstrate such a relation, also in prelingually deafened adults (Busby & Clark, 2000; Gifford et al., 2018). The presence of such a relation suggests that an optimization of the coding of spectral cues could have a positive impact on speech understanding outcomes as well. The success of studies, including our own, attempting to improve spectral resolution by means of deactivating electrodes that poorly encode speech information, is however mixed (Henshall & McKay, 2001; Saleh et al., 2013; Vickers, Degun, Canas, Stainsby, & Vanpoucke, 2016; Zwolan et al., 1997).

A recent study by Sagi and Svirsky (2018) offers new explanations for why purely removing the poorest encoding electrodes from the fitting map might not be the best solution. By means of extensive mathematical modelling, they were able to demonstrate that the main limiting factor in previous studies is the fact that the criteria used to deactivate the poorest encoding electrodes do not take the distribution of speech information over the remaining electrodes into account. Their results show that the benefit in speech recognition was significantly greater when an electrode deactivation pattern was used that maximizes the discrimination of speech cues ("model-optimized"), compared to a deactivation pattern that simply removes the poorest encoding electrodes ("best electrodes"). For the poorer performing group in this study (electrode discrimination difference limens between 0 and 4 electrodes), which is most representative for our study

group of early-deafened CI users, maximum improvements were demonstrated for the model-optimized approach when 11 out of the 22 electrodes remained active.
A recurring bottleneck, both in our study (*chapter 3*) as in the model of Sagi and Svirsky (2018), is that complete adaptation to each new combination of electrodes is required in order to attain a maximal score for this combination. Presuming however, that subjects are able to completely adapt given sufficient time, these new insights demonstrate that combining a measure for identifying poorly discriminating electrodes with a model approach focusing on optimally transferring speech information, has significant potential for improving outcome. This is an interesting line of research which requires further exploration in our target group, although the time-consuming measurements might be a practical obstacle to find sufficient numbers of subjects willing to participate.

Measuring outcome: a different perspective

In *chapters 2 and 3* we focused on how technical aspects of CI processing are related to speech understanding outcomes in the group of early-deafened CI users. In *chapter 6* however, one of the aims was to explore how we should measure outcome in our target group in the first place. Traditional outcome measures for CI users mainly include open-set word and sentence recognition tests in quiet, evolving towards measures in noise as performance of the average, postlingual CI user improves, as illustrated by the recommendations of the New Minimum Speech Test Battery (MSTB) for adult cochlear implant users (Auditory Potential, 2011). This shift towards more complex outcome measures tends to forget that a significant proportion of adult CI users, especially in the early-deafened group, still struggles to obtain any open-set speech recognition benefit at all (Caposecco et al., 2012; Heywood et al., 2016; O'Gara et al., 2016; Rousset et al., 2016).
In *chapter 6* our subject group of 27 early-deafened subjects obtained a mean 1 year post-operative CVC word recognition score of 44.2%, which corresponds with the range of outcomes (20-49%) of the studies reviewed in *chapter 4*. Out of the 27 subjects in our subject group, however, 7 obtained no more than 10% and 10 subjects scored less than 30% on this open-set CVC word test. This latter group of 10, the so-called “poor performers”, did show significant benefit on a closed-set measure of suprasegmental cues as well as on a measure of speech tracking, assessing the general ease of communication.
Part 1 of our systematic review (*chapter 4*) equally found measures of closed-set speech understanding to be valuable and although there were no studies included that assessed speech tracking, there is a similarity with tests assessing auditory-visual communication. The latter tests showed significant benefits in almost all studies, pointing to the capability of early-deafened subjects to combine new, auditory information from the CI with visual information obtained through lip reading, thus obtaining a higher level of speech understanding. Our recommendation therefore is to substitute traditional outcome measures of speech understanding with closed-set measures and measures assessing the

combination of auditory and visual information in this patient population, especially in poor performers. To this end, we are currently working on an extended validation of the (Dutch) texts used for the speech tracking test we implemented in this population, in order to improve their clinical usefulness.
In addition, it was shown in *chapter 6* that the early-deafened subject group, but also the subgroup of poor performers, showed significant benefit on the questionnaire on subjective benefit, the latter despite the absence of a significant improvement on open-set speech understanding. Very positive outcomes for hearing-related quality of life were confirmed in *chapter 4*. In general, changes in subjective experience after cochlear implantation were most clear for domains related to sound perception, sense of safety and communication, but were less obvious with respect to social life or self-esteem, which we believe might be due to the long-standing nature of the subjects' hearing impairment. The questionnaire used in *chapter 6*, which was specifically developed to be used in our target population, fulfils the need for a questionnaire that takes the particularities of this patient group into account. On the other hand, the limited number of subjects it was submitted to unfortunately did not allow for an extensive validation. This is an issue that should be further addressed prior to a wider use of the questionnaire. Finally, the lack of a strong relation between subjective benefit on one hand and speech perception outcome on the other, emphasizes that a subjective measure of outcome is indispensable in our target population to obtain a truthful assessment of outcome.

Predicting performance

The second part of the systematic review presented in *chapter 5* identified three variables as having good potential in predicting (speech understanding) outcomes in the early-deafened population; one of those, the preoperative word recognition score, was one of two remaining significant predictors in the multiple regression analysis performed on our own study group as well (*chapter 6)*. The two other variables that were identified as good predictors in *chapter 5*, i.e. preoperative speech intelligibility and communication mode in childhood, were not incorporated in our own analysis. This can mainly be attributed to a lack of data on these variables as they are not systematically assessed in our CI-candidates. Inversely the second significant predictor of our own regression analysis, pre-operative hearing thresholds of the ear to be implanted, was not clearly related to outcomes in the studies of the review; the thresholds of the better ear showed more predictive potential. The small number of subjects our multiple regression analysis was based on inevitably limits the generalizability of the observed outcomes. There is clearly a need for research in larger groups of early-deafened subjects, which at the same presents a major challenge given the relatively small proportions of early-deafened CI users implanted in most clinics. A multicentre study, as has been performed in postlingually deafened CI users (Blamey et al., 2013), could potentially provide an answer to this problem.

Apart from sample size issues, the major limitations observed in *chapter 5* which should be taken into account by future studies, were the lack of clear definitions of the included variables, as well as inappropriate statistical analysis. Attention also needs to be paid to the choice of predictors to include. Until now, the focus of most studies - including our own - has been on demographic factors concerning hearing history and the implantation itself. The combination of these factors has never been be able to explain much more than about 60% of the variation in speech understanding outcome in this population, with a great deal of variables mainly explaining the same variation (*chapter 6*, Caposecco et al., 2012; Kraaijenga et al., 2016; O'Gara et al., 2016; van Dijkhuizen et al., 2011).

Recently, Pisoni et al. (2017) suggested that the missing predictors should be sought in measures of underlying cognitive processing, at least in postlingually deaf adults. In that respect, Kral et al. (2016) suggest that the limited auditory experience during the development of early-deafened subjects not only disturbs the perception of spoken language and development of the auditory system itself, but also affects the development of neurocognitive functions such as concept formation and executive functioning, and results in altered connections between the auditory system and other brain systems. Individual variability in the development of neural circuits in response to auditory deprivation is thought to contribute to the observed variability in outcomes with a cochlear implant later on. Subjects, who are for instance better at integrating top-down information streams with the incoming auditory cues, are more likely to become better performers. These ideas can be extended to the population of late-implanted but early-deafened CI users: the extent to which these higher-order neurocognitive functions have been developed might be determined by the amount of auditory input in early childhood (which is related to the exact age at onset of deafness, the amount of residual hearing and the adequacy of early hearing aid rehabilitation), along with the aforementioned individual variability. It would thus be an interesting area for further investigations to assess if and which measures of neurocognitive functioning could be used within our target population, and whether they are valuable as predictors in multiple regression analyses.

Finally, when taking in mind that a measure for auditory-visual speech recognition is often a more relevant way to assess outcome in early-deafened CI users (*chapter 4 and 6)*, it would equally be interesting for future studies in our target population to find relevant predictors for such an auditory-visual outcome measure, instead of just focusing on auditory-only benefits. As the ability to integrate these communication modes potentially requires different skills, the relevant predictors might differ as well.

Reflecting on implantation criteria

Although cochlear implantation in patients with early-onset deafness remains controversial within the Deaf community, especially when it concerns young children (Sparrow, 2010), it was not within the scope of this thesis to go further into this debate. With respect to early-deafened adults we believe that it is a personal choice for every individual to sign up, or not, for cochlear implantation. As the technique of cochlear implantation was not yet applied on a large scale in the Netherlands until the late 90's, parents of children born deaf or severely hearing impaired before that time did not have the possibility to choose for a cochlear implant, as is the case nowadays.

The results presented in *chapters 4, 5 and 6* urge us to reflect on the implantation criteria for those early-deafened adults who are interested in increasing their access to the hearing world and are now candidates for cochlear implantation. A very interesting observation was that the evidence so far does not strongly support the notion that subjects showing large changes in hearing-related quality of life are also the ones obtaining significant speech understanding improvements. Although more research is definitely required, a strong correlation between auditory gains and subjective benefits could be found neither in our study group nor in the review. In addition, the anticipated gain in auditory-only speech understanding with CI is generally limited, whereas it was also demonstrated that when subjects are evaluated with assessment methods targeted to their level of auditory functioning, benefits often do arise, but they might be on domains other than open-set speech understanding.

Based on these observations, we suggest that implantation criteria in this population should not only look at the expected level of auditory-only speech understanding, but also include alternative domains of potential benefit including auditory-visual speech recognition and hearing-related quality of life.